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Unilateral proptosis

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SUMMARY Twenty-seven children with unilateral proptosis were investigated over a period of 8½ years. Seventeen (63%) of them had local or disseminated malignancy, a higher proportion than in earlier reports. The frequency of inflammatory lesions, particularly ethmoiditis progressing to proptosis, appears to have decreased, probably because of the liberal use of antibiotics for upper respiratory tract infections.

Proptosis is a rare presenting complaint in childhood. Analysis of the causes in children investigated since 1970 at Birmingham Children's Hospital and at Birmingham and Midland Eye Hospital shows a change, compared with earlier series,[1–3] in the importance of malignant and inflammatory diseases. An investigative protocol is suggested which takes into account this change and the availability of new diagnostic techniques.

The series includes causes of proptosis not previously reported in children.

Materials and methods

We reviewed the medical records of all children attending these two Birmingham hospitals for the investigation of proptosis between January 1970 and July 1979. We excluded any child with bilateral exophthalmos, pseudoproptosis (for example monocular buphthalmos or unilateral high myopia), and obvious cranio-facial anomalies in which proptosis was a feature.

Results

Twenty-seven children were investigated during the 8½ years. The final diagnoses are shown in Tables 1 and 2 and it can be seen that 17 (63%) cases resulted from entirely local intraorbital disease.

Local causes of unilateral proptosis (Table 1).

Developmental

Developmental lesions were responsible for proptosis in 5 children, 4 of whom presented in the first year of life.

A computerised tomography (CT) scan had been performed on Case 5 (Fig. 1) and interpreted as showing an optic nerve glioma; later, on orbital exploration, this proved to be a blood cyst.

Table 1 *Aetiology of unilateral proptosis in children—local causes*

Case	*Age at presentation*	*Diagnosis*
Developmental lesions		
1	20 days	Lymphangioma
2	7 months	Haemangioma
3	4 months	Haemangioma
4	24 days	Encephalocoele
5	3 years	Blood cyst
Neoplastic lesions		
6	13 months	Glioma
7	11 years	Glioma
8	6 years	Rhabdomyosarcoma
9	28 months	Rhabdomyosarcoma
10	29 months	Rhabdomyosarcoma
11	5 years	Rhabdomyosarcoma
12	10 years	Rhabdomyosarcoma
13	11 years	Fibrosarcoma
14	16 months	Retinoblastoma
Inflammatory lesions		
15	4½ years	Cavernous sinus abscess
16	15 months	Ethmoiditis
17	3 months	Osteitis

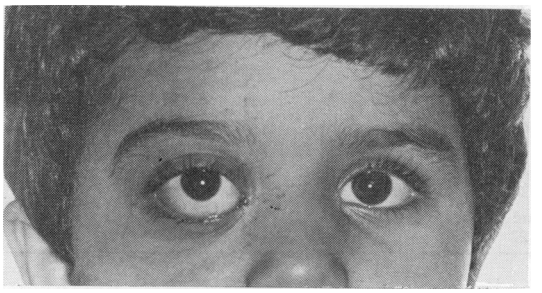

Fig. 1 (*Case 5.*) *Right axial proptosis.*

Neoplasia

A neoplastic lesion was the cause in 9 of the 27 children. Five had rhabdomyosarcoma, 4 presenting with firm lid swelling before the onset of proptosis. The histological diagnosis was obtained by biopsy of the lid lesion in 2 patients.

In addition 2 children had optic nerve gliomas,

one had fibrosarcoma, and one an orbital extension of a retinoblastoma.

Inflammatory
There were only 3 patients with local inflammation; in 2 this was a result of ethmoiditis, and in 1 it was due to cavernous sinus abscess arising 3 months after orbital trauma.

Systemic causes of unilateral proptosis (Table 2).

Developmental
The only child in this group (Case 18) had a variant of the McCune-Albright syndrome. A CT scan is shown in Fig. 2.

Neoplasia
There were 6 children with disseminated malignant disease which presented with proptosis. Acute leukaemia was the most common cause, 1 child having acute myeloid, 1 myelomonocytic, and 1 acute lymphoblastic leukaemia. Two children had neuroblastoma and one 11-year-old girl had histiocytosis.

Physical examination led us to suspect a generalised disease in all of these children, most often because of anaemia, and, in 4 children, because of hepatosplenomegaly.

Table 2 *Aetiology of unilateral proptosis in children—systemic causes*

Case	*Age at presentation*	*Diagnosis*
Developmental lesions		
18	7 years	McCune-Albright syndrome
Neoplastic lesions		
19	7 years	Neuroblastoma
20	10 months	Neuroblastoma
21	18 months	Acute myeloid leukaemia
22	18 months	Acute myelomonocytic leukaemia
23	6 years	Acute lymphoblastic leukaemia
24	11 years	Histiocytosis
Inflammatory lesions		
25	11 years	Tuberculosis
Endocrine		
26	11 years	Graves's disease
Undiagnosed		
27	8 years	

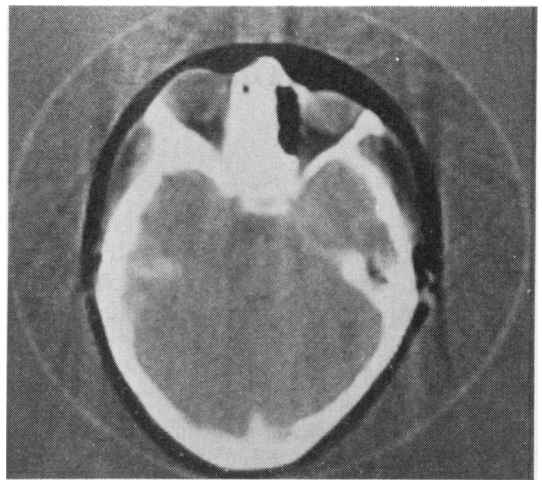

Fig. 2 *(Case 18.) Computerised tomography showing thickening of the left sphenoidal wing, ethmoid and maxillary antrum, with subsequent reduction in volume of the left orbit.*

Inflammatory
The girl in this group had an orbital tuberculoma.

Thyroid diseases
A 12-year-old girl had normal thyroid function tests at presentation with proptosis, but developed florid Graves's disease four years later.

Discussion

This review reinforces our initial impression that the disease underlying unilateral proptosis is changing. A series reported in 1967 contained only 16% of neoplastic cases (Table 3). In 1972, a second study recorded an incidence of 42%, and in the present series neoplasia accounted for 63%. A report of 65 children from Mexico[2] (Table 3) was not considered representative because of the high number of cases of advanced retinoblastomas, rarely seen in Britain.

In contrast, only 4 (15%) of our patients had an underlying inflammatory lesion compared with 30% reported in 1967.[1] This change may indicate that the earlier and more widespread use of

Table 3 *Comparison of reported paediatric series*

Place	*Authors*	*Year*	*Number of patients*	*Local causes*	*Aetiology*	
					Inflammatory	*Neoplasia*
Hospital for Sick Children, Toronto, Canada	Ophthalmological Staff of the Hospital for Sick Children, Toronto, Canada.[1]	1967	257	61%	30%	16%
Mexico City, Mexico	Silva[2]	1968	65	93%	17%	51%
Lebanon	Zakharia *et al.*[3]	1972	21	81%	45%	42%
Birmingham, UK	Oakhill *et al.*	1981	27	63%	15%	55%

antibiotics for upper respiratory tract infections prevents ethmoiditis. The series in Toronto was studied during a period that included a time before the introduction of antibiotics. Examination of the individual case histories stresses certain clinical points which are worthy of elaboration. For example, lid swelling may indicate rhabdomyosarcoma of the orbit and was present in 4 of our 5 cases, and in 25 of 29 in a previous report.[4] Early diagnosis may be possible from biopsy of the lid lesion without resorting to orbitotomy. Orbital blood cysts are rare and may be the cause of a superior orbital fissure syndrome in conjunction with proptosis.[5] They probably arise from haemorrhage into a pre-existing hamartomatous malformation and are not simply the result of trauma. The particular significance of the child described here (Case 5) is that the results of sophisticated investigative techniques must be interpreted after careful clinical assessment. In this patient the history of rapid onset, coupled with normal vision, made optic nerve glioma an unlikely diagnosis despite all radiological features.

Monostotic fibrous dysplasia is not generally associated with precocious puberty, but it did lead to unilateral exophthalmos in 7 of 86 patients reported by Van Buren[6] and in 6 (2 children) of 300 in Silva's series.[2] Our patient is unusual because she has monostotic fibrous dysplasia and precocious puberty, but not the other features of the McCune-Albright syndrome (polyostotic fibrous dysplasia and skin pigmentation). Daves and Yardley[7] suggested that monostotic fibrous dysplasia is related to the McCune-Albright syndrome and our patient's condition seems to support their hypothesis.

Disseminated malignancy was the most common type of generalised disease to cause proptosis in our series. Acute leukaemia and neuroblastoma are noted for presenting in this way. For example, Blake and Fitzpatrick[8] reported that 8 of 15 cases of neuroblastoma had orbital metastases at some stage.

Tuberculosis affecting the orbit, which is very rare, has not been reported in childhood. In adults it is generally haematogenous in origin and may present with proptosis,[9] orbital apex syndrome, or a superior orbital fissure syndrome.[5] Diagnosis was made easier in our patient by a positive family history and the ease of biopsy from a femoral lesion.

Thyroid eye disease is responsible for 16% of unilateral proptosis in adults[10] but is a rare cause in children. Although many investigative techniques are available for the assessment of unilateral proptosis, we consider that the following investigations are useful in leading to a rapid and accurate diagnosis: (1) If, after clinical examination, the disease appears to be intraorbital then skull *x*-ray films, CT scan, and ultrasound will be useful before biopsy.
(2) If the disease appears to be systemic, in addition to the investigations suggested for local causes, full blood count, bone marrow aspirate, *x*-ray films of chest, abdomen, and skeleton, and 24-hour urine collection for catecholamines and thyroid function tests should be performed.

Price and Danziger[11] reported the value of CT scanning in the assessment of orbital tumours in children, and Dallow,[12] using a combination of CT and ultrasound, claimed he was able to make the correct diagnosis in 98% of his adult patients. These two non-invasive techniques have become of prime importance in the investigation and follow-up of proptosis.

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